

# **AN ANALYSIS OF RESULTS OF DEROTATION OSTEOTOMY THROUGH THE SYNOSTOSIS MASS IN CASES OF CONGENITAL RADIOULNAR SYNOSTOSIS**

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**The Tamilnadu Dr.M.G.R.Medical University**

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## **CERTIFICATE**

This is to certify that this dissertation titled “**AN ANALYSIS OF RESULTS OF DEROTATION OSTEOTOMY THROUGH THE SYNOSTOSIS MASS IN CASES OF CONGENITAL RADIOULNAR SYNOSTOSIS.**” submitted by **DR.S.SIVA SWAMINATHAN** to the faculty of Orthopaedics, The Tamilnadu Dr. M.G.R. Medical University, Chennai in partial fulfillment of the requirement for the award of MS degree ( branch – II) Orthopaedics, is a bonafide research work carried out by him under our direct supervision and guidance.

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## **DECLARATION**

I, **DR. S.SIVA SWAMINATHAN**, solemnly declare that the dissertation titled “**AN ANALYSIS OF RESULTS OF DEROTATION OSTEOTOMY THROUGH THE SYNOSTOSIS MASS IN CASES OF CONGENITAL RADIOULNAR SYNOSTOSIS**” has been prepared by me. This is submitted to **The Tamilnadu Dr. M.G.R. Medical University, Chennai**, in partial fulfillment of the regulations for the award of MS Degree ( Branch II) Orthopaedics

**Place: Madurai**

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**Date:**

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## **INTRODUCTION**

Congenital radioulnar synostosis is a rare congenital anomaly when the failure of segmentation of radius and ulna results in a fixed position of the forearm ranging from neutral rotation to severe pronation(1).When the deformity is mild, little disability is evident, as the ipsilateral shoulder and wrist can compensate effectively but with significant pronation the activities of daily living including dressing, eating, grasping objects in the palm of the hand can be severely impaired.

Attempts at resection of the synostosis in order to restore rotation of forearm have, on the whole been unsatisfactory due to loss of correction and vascular compromise following extensive soft tissue release (2,3,4,5,6). More recently osteotomy through the synostosis mass and fixing the forearm in a functional position has been advocated.

In this study we describe the surgical technique of osteotomy through the synostosis mass, results and advantages of this procedure over other surgeries.

## **AIMS AND OBJECTIVES**

1. To evaluate the results and functional outcome of correcting the deformity by an osteotomy through the synostosis mass in cases of congenital proximal radioulnar synostosis.
2. To study the advantage of this procedure over other techniques of osteotomy.



## **HISTORICAL BACKGROUND OF CONGENITAL RADIOULNAR SYNOSTOSIS**

Congenital Radioulnar Synostosis is an uncommon deformity of the upper extremity. Blaine (7), Mauchet and Leleu (8) stated that radioulnar stenosis was first discovered by Lenoir at an autopsy in 1817. However, the earliest record is that of Sandifort, who, in 1793, reported three cases(9). Smith, Verneuil and Dubois reported a single case in 1852. In 1856, Malgaigne wrote about one case and was followed in 1863 by Voigt.



In 1892, Morrison reported one case and Abbott contributed to the largest series of cases, comprising of 7 cases. In 1932, Fahlstorm reviewed all of the reported cases in the world literature and found only 185 since Sandifort's original description in 1793 (9).

In 1924, Davenport (10) reported the largest series of 15 cases and covered this condition more thoroughly. Davenport, as the major essayist of this group, made a complete study regarding congenital radioulnar synostosis.

In order to have a thorough understanding on congenital radioulnar synostosis, one should know the developmental embryology of upper limb.

## **EMBRYOLOGY-DEVELOPMENT OF UPPER LIMB**

The embryogenesis of the upper extremity commences with the formation of the upper limb bud on the lateral wall of the embryo four weeks after fertilization. The developing limb bud consists of a mesenchymal core that is covered by epithelial tissue of ectodermal origin (11). Three signaling centers that control different aspects of limb development have been discovered (12,13,14,15).

1. APICAL ECTODERMAL RIDGE (AER)
2. ZONE OF POLARISING ACTIVITY (ZPA)
3. WINGLESS-TYPE (Wnt) SIGNALLING CENTER

The apical ectodermal ridge is a layer of ectoderm that condenses over the limb bud and acts as signaling center to guide the underlying mesoderm to differentiate into appropriate structures (12,15). It is obligated for limb development from proximal to distal direction and is responsible for interdigital necrosis, which separates the webbed hand.

The proliferating mesenchyme in the area subjacent to the AER is known as the PROGRESS ZONE (16). The subpopulation of cells at the posterior border of the Progress zone is the ZPA (17) which functions as a signaling center for the anterior-to-posterior (radioulnar) limb development (12,18).

The Wnt signaling center resides in the dorsal ectoderm and secretes factors that induce the underlying mesoderm to adopt dorsal characteristics. It is responsible for development of dorsal to ventral axis configuration and the alignment of the limb with a dorsal orientation (dorsalisation) (19).

The AER, ZPA and Wnt pathway all function in a co-ordinated effort to ensure proper limb patterning and growth during embryogenesis (12).

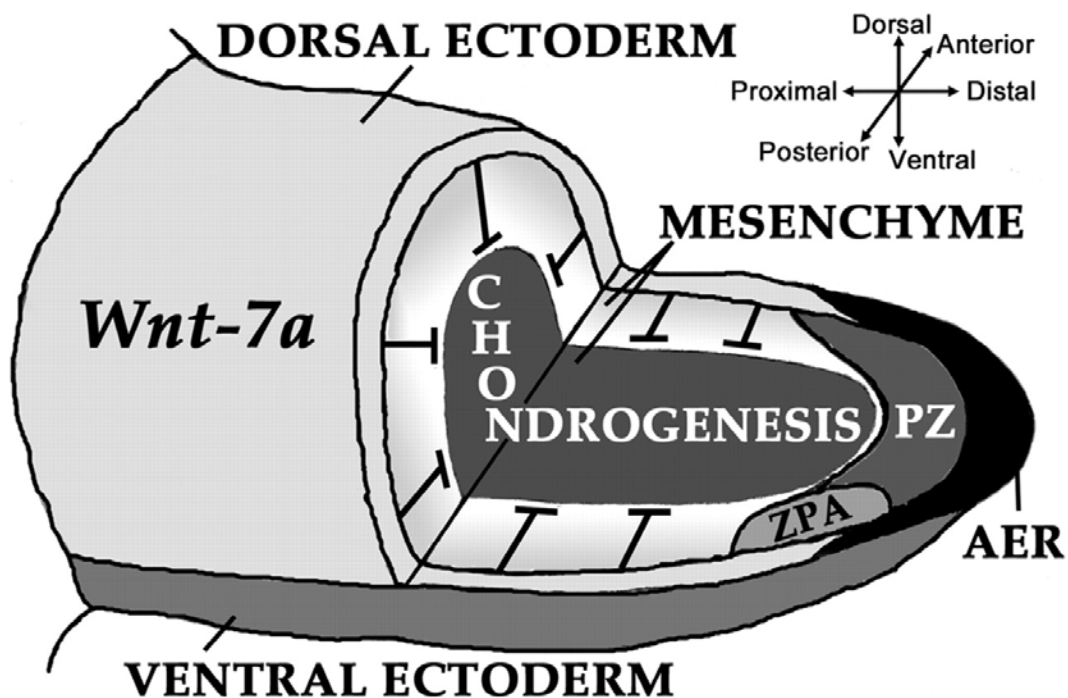
Abnormalities within one signaling center indirectly prohibit adequate functioning of other two remaining centers and affect limb formation.

## SIGNALING CENTER PATHWAYS DURING EMBRYOGENESIS

Signaling center	Responsible substance	Action
Apical Ectodermal Ridge	Fibroblast growth factor (FGF)	Proximal to distal limb development, interdigital necrosis
Zone of Polarizing Activity	Sonic hedgehog protein (SHH)	Radioulnar(anteroposterior)formation
Wnt pathway	-	Dorsalisation of limb

The substance for proper functioning of AER is FGF and that of ZPA is SHH protein.

### SIGNALLING CENTERS

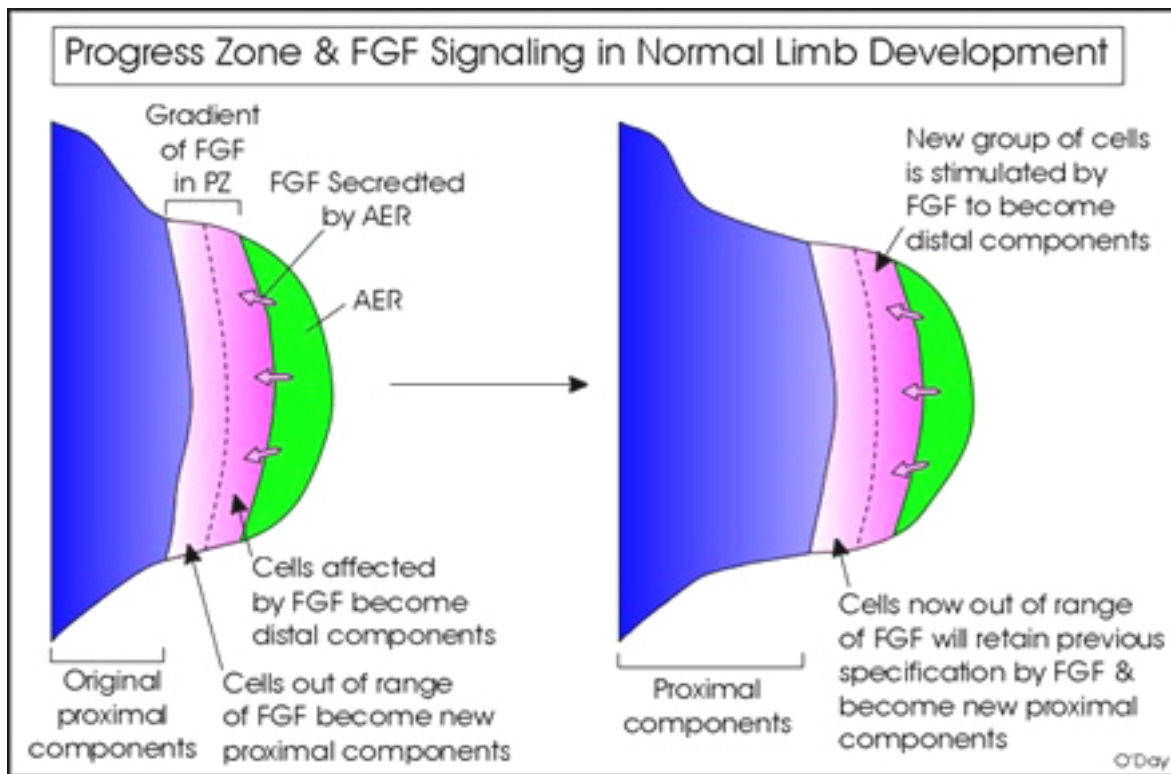


## OUTGROWTH AND PROXIMODISTAL PATTERNING

AER is the center responsible for proximodistal patterning. As the limb bud grows out, the limb structures are laid down in sequence, starting with proximal structures and finishing with distal structures such as the digits. The FGF's are the mediators of AER activity.

## ANTEROPOSTERIOR (RADIOULNAR) PATTERNING

The anteroposterior patterning of the limb is specified by a signal (SHH protein) from the ZONE OF POLARISING ACTIVITY at the posterior margin of the limb bud (20). The interaction between the polarizing region and the cells of the progress zone ensures that, for example, the correct sequence of digits develop, with the thumb at one edge of the hand and the little finger at the other.



One model of polarizing region signaling proposes that the polarizing region produces a diffusible morphogen that sets up a concentration gradient across the limb bud (21). Cells at different distances from the polarizing region would be exposed to different concentration of morphogen and the local morphogen concentration would control the digit identity.

## DORSOVENTRAL PATTERNING

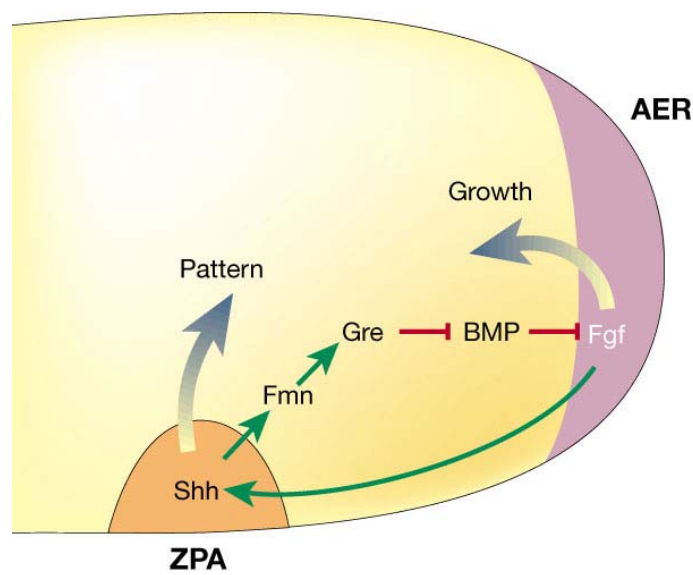
The function of dorsoventral patterning of the limb is performed by the Wnt pathway which resides in the dorsal ectoderm.

## MOLECULAR BASIS OF SIGNALLING IN THE DEVELOPING LIMB

### OUTGROWTH SIGNALS

The initial outgrowth of the limb is stimulated by FGF 8. Once the limb bud is established along with FGF 8, FGF 4 and FGF 2 are expressed in the AER and are required for normal limb development. Specific function of FGF seems to be that of maintenance of the PROGRESS ZONE thereby aiding in continued outgrowth of limb bud. In addition to FGF, transcripts of several genes are found in the AER viz., genes encoding for Bone Morphogenetic Proteins (BMP's). Transcripts of the *ld* gene (limb deformity gene) are also found in the AER. A novel set of proteins, FORMINS, which are localized in the nucleus are encoded by the *ld* gene.

### MOLECULAR BASIS OF SIGNALLING



## POLARIZING SIGNALS

Two molecules can reproduce signaling of the polarizing region Retinoic acid (22) and Sonic Hedgehog gene (18).

These putative signaling molecules have quite different properties; retinoic acid is highly diffusible in limb whereas the protein encoded by sonic hedgehog acts locally.

A bone morphogenetic protein, BMP 2 is also expressed in the polarizing region (23). BMP-4 is expressed in both anterior and posterior mesenchyme at early stages. BMP-2 expression in anterior cells is activated by retinoic acid suggesting that BMP-2 plays a role in A-P patterning.

The expression of FGF's in the AER is regulated by SHH. Once induced, SHH expression in AER can be maintained by FGF-4 creating a positive feedback loop between ZPA and AER (24). This feedback loop through which outgrowth and patterning along the A-P, proximal to distal axes of the developing limb bud occurs in an orderly regulated fashion sustained by SHH and FGF-4 expression.



## DORSOVENTRAL SIGNALS

Transcripts of Wnt-7A are localized strictly to the dorsal ectoderm and are not found in the ventral ectoderm (25, 26). Wnt-7A is therefore a good candidate for providing dorsal ectoderm signal.

## CLASSIFICATION OF LIMB ANOMALIES

The history of classification of congenital limb anomalies is rich. Sainte Hilaire initially classified these 'vices of conformation' as slight or severe in 1829 (27).

In 1831, Otto (28) grouped these 'vices of organization' according to ten variations (number, size, form, position, connection, colour, consistency, continuity, texture and content).

In 1832, Sainte Hilaire (29) coined the terms ectromelia, hemimelia and phocomelia. Leboucq (30) noted the longitudinal arrangement defects.

Two important contributions from the German literature were made by Kummel (31) in 1895 and later modified by Nigst (32) in 1927, who divided congenital anomalies into three categories

- 1) Defect malformation (i.e., deficiencies)
- 2) Syndactyly (i.e., fusion of parts)

### 3) Polydactyly (i.e., excessive number of parts)

The current classification scheme was first proposed by Swanson in 1964 (33, 34) and was based on the premise that anomalies should be grouped according to parts of the limb that have been primarily affected during development.

There were six basic categories in the initial proposal: failure of differentiation of parts, arrest of development, duplications, overgrowth, congenital circular constriction bands and generalized skeletal defects. The IFSSH and ISPO then added a seventh basic category, undergrowth.

## INTERNATIONAL FEDERATION OF SOCIETIES FOR SURGERY OF THE HAND (IFSSH) CLASSIFICATION OF LIMB ANOMALIES

### 1) FAILURE OF FORMATION OF PARTS

#### A. Transverse deficiencies

#### B. Longitudinal deficiencies

##### 1. Phocomelia

##### 2. Radial

##### 3. Central

##### 4. Ulnar

## 2)FAILURE OF DIFFERENTIATION

A. Synostosis

B. Radial head dislocation

C. Symphalangism

D. Syndactyly

E. Contracture

### 1. Soft tissue

a) Arthrogryposis

b) Pterygium

c) Trigger

d) Absent extensor tendons

e) Hypoplastic thumb

f) Clasped thumb

g) Retroflexible thumb

h) Campodactyly

i) Windblown hand

### 2. Skeletal

a) Clinodactyly

b) Kirner deformity

c) Delta bone

### 3) DUPLICATION

1. Thumb
2. Triphalangism/hyperphalangism
3. Polydactyly
4. Mirror hand

### 4) OVERGROWTH

1. Limb
2. Macrodactyly

### 5) UNDERGROWTH

### 6) CONGENITAL CONSTRICTION BAND SYNDROME

### 7) GENERALISED SKELETAL ABNORMALITIES

## CONGENITAL RADIOULNAR SYNOSTOSIS

Congenital radioulnar synostosis comes under ‘failure of differentiation of parts’ according to the above classification. Although the condition is present since birth, a delay in presentation is common until the child begins engaging in more complex daily activities. The functional implication of a congenital fusion depends on the motion normally present between the two bones and the ability of the adjacent joints to compensate.

## PATHOANATOMY

Lewis described the humerus, radius and ulna as being continuous with each other, and joined by a common perichondrium, at five weeks of gestation. By six weeks, the cartilaginous anlage of the three bones is separated by condensation of tissue and no joint cavities are yet visible. The forearm is in a neutral position at this time, although rotation into pronation occurs by eight weeks due to growth discrepancy between the arterial tree and the radius (35).

It is the failure of differentiation and the persistence of the cartilaginous anlage between the radius and ulna during the seventh week of development that results in a persistent bridge of tissue (36). Usually this will ossify into a osseous synostosis , although fibrous synostoses are also well recognized (6,10,37). A fixed, pronated forearm is thought to reflect the developmental arrest at this specific time of fetal development. The frequently associated deformity of radial head may be due to early interference with joint formation that results in a complete proximal coalition (6) or to limited fusion distal to the epiphysis that results in unequal growth of the radial head.

## EPIDEMIOLOGY

Congenital radioulnar synostosis is a rare congenital anomaly. The rarity of this condition often leads to delayed clinical diagnosis. Cleary and Omer reported an average patient age at diagnosis of 6 years with a range from 6 months to 22 years (38). There is no sex predilection. Sixty percent of cases are bilateral.

## CLINICAL FEATURES

Although the condition is present since birth, it is usually not evident until early adolescence when the patient presents with lack of supination and pronation of the forearm. Functional deficits associated with congenital radioulnar synostosis depend on the severity of the deformity and whether or not it is bilateral.

Congenital radioulnar synostosis is a deformity characterized by a fixed position of the forearm ranging from neutral rotation to maximum pronation. The position is usually one of pronation or hyperpronation. More than 60% of cases are bilateral(9). Males and females are affected in similar numbers(3).



**Picture showing difficulty in buttoning shirt in a patient with congenital radioulnar synostosis on the left side**

There is usually full or near full range of elbow motion. Elbow flexion contracture rarely exceeds 30 degrees. There may be an abnormal carrying angle at the elbow and shortening of the forearm. Pain is uncommon (39). Hypermobility at the radiocarpal and midcarpal joints often disguises these forearms lacking rotation, particularly when the fixed position is in neutral rotation or mid pronation(40).

## DISABILITY

A child with severe forearm deformity usually has significant functional limitation when they try to hold a rice bowl, drink water from a glass or receive a coin in the open palm and they have inability to use spoons and wash their face.



**Backhand position of left upper limb causing difficulty in bringing hand to  
mouth**



## PATIENT ADAPTATION

In a forearm fixed in supination, pronation is achieved by internal rotation of shoulder, elevation of the elbow and intercarpal pronation. With this maneuver, the hand can complete activities such as eating and writing, although performing these activities with the elbow high in the air may be cosmetically objectionable. In a forearm fixed in pronation, compensation for supination by external rotation of the shoulder and the intercarpal supination is insufficient to place the pronated forearm in supinated position. When pronation is marked, the inability to supinate to a neutral position can make activities such as drinking from glass difficult or impossible.

## ASSOCIATED ANOMALIES

Since Congenital radioulnar synostosis is caused by an in utero insult, its association with other abnormalities is not surprising. About one third are associated with general skeletal abnormalities, such as hip dislocations, knee anomalies, club foot, polydactyly, syndactyly, madelung deformity, microcephaly, multiple exostoses, ligamentous laxity, thumb hypoplasia, carpal coalition and other cardiac, renal and gastro intestinal tract abnormalities (6,36,39).

## SYNDROMIC ASSOCIATION

Simmons et al documented one third of cases with associated abnormalities, some of which are genetically determined. Associated syndromes include Apert syndrome, Carpenter syndrome, Arthrogryposis Multiplex Congenita, Mandibulofacial dysostosis, Williams syndrome, Klinefelter syndrome, Holt-Oram syndrome, Fetal Alcohol syndrome and Poland syndrome (41,42).

## FAMILIAL RADIOULNAR SYNOSTOSIS

The first report of familial radioulnar synostosis was by Abbott in 1892 and approximately 18 families have been described till date. Almost all of these families are of Western European origin, several of Jewish descent.

Familial radioulnar synostosis tends to be bilateral and a majority of the cases are male. The inheritance pattern is autosomal dominant with incomplete penetrance.

## RADIOLOGICAL FINDINGS AND CLASSIFICATION SYSTEMS

Routine anteroposterior and lateral radiographs of the involved elbow joint with forearm would be sufficient to confirm the diagnosis. A CT scan may be helpful to know the exact extent of the synostosis mass and to classify the

condition. There are a number of classification systems based upon the radiographic findings.

Wilkie classified the synostosis roentgenographically into two types (43).

TYPE 1:

There is a complete synostosis with the radius and ulna fused at their proximal borders for a variable distance.

TYPE 2:

The synostosis is just distal to the proximal radial epiphysis, is more likely to be partial, and is associated with dislocation of the radial head.

Simmons et al preferred to consider the malformation as a spectrum of anomalies with varying degrees of length of the synostosis, with or without involvement of the radial head (6).

Mittal classified congenital radioulnar synostosis into two types (36).

TYPE 1:

Proximal radial epiphysis and metaphysis fused to the ulna

**TYPE 2:**

Fusion distal to proximal radial epiphysis and is associated with dislocation of the radial head.

Cleary and Omer classified Radioulnar synostosis into four types based on the radiographic appearance (38).

**TYPE 1:**

Clinical evidence of radioulnar fusion with a normal radiograph.

**TYPE 2:**

Bony radioulnar synostosis with normal radial head.

**TYPE 3:**

Bony radioulnar synostosis with hypoplastic posteriorly dislocated radial head.

**TYPE 4:**

Short synostosis with mushroom shaped radial head dislocated anteriorly.

## MANAGEMENT

The disability encountered by patients with congenital radioulnar synostosis is usually insignificant when the forearm is fixed in supination, neutral rotation or mild pronation. The patient would be able to perform the activities of daily living by compensatory ipsilateral shoulder motion and wrist hypermobility.

## INDICATIONS FOR SURGERY

The indication for operation for treatment of congenital radioulnar synostosis is controversial. Cleary and Omer believe that the operative intervention is rarely indicated and their surgical indication is based more on functional deficits than absolute forearm position (38). On the other hand, Simmons and Southmayd concluded that an operation is indicated in a patient with greater than 60 degrees of fixed pronation and must be individualized in patients with degrees of pronation between 15 to 60 degrees (6). Ogino and Hikino also reported that patients with greater than 60 degrees of pronation had restrictions in daily activities. Generally, surgical intervention is dependent on functional limitations, type of involvement (bilateral or unilateral, dominant or non dominant side), social and cultural environment of the patient, and projected future activities (40).

## TIMING OF SURGERY

Griffet et al recommended that the surgery is best carried out between the ages of 4 and 10 years (44). At these ages, the osteotomy is easy, and it is likely to ensure sufficient remodeling of the radius and ulna. Also vascular and neural postoperative complications were higher in older age group.

## GOALS OF SURGERY

- 1) To restore forearm rotation (supination and pronation) or to bring the forearm to a functional position.
- 2) To prevent the recurrence of bony ankylosis between the radius and ulna.

## SURGICAL TREATMENT OPTIONS

Attempts to obtain and maintain rotational motion of the forearm have been for the most part unsuccessful with reformation of the synostosis in spite of excision. Several operative procedures have been suggested for congenital radioulnar synostosis, that can be grouped into two major groups.

- 1) Operations that were designed to restore the rotational motion of the forearm (i.e., supination and pronation) in addition to removal of the synostosis.
  - a) Interposition of swivel prosthesis (Kelikian and Doumanian)

b) Osteotomy and interposition of soft tissues- silastic membrane, anconeus and free vascularised fascio-fat graft.

2) Operations that improve the fixed position of the forearm into a more functional position. Various types of derotation osteotomy have been described in this regard.

a) Osteotomy at the synostosis mass.

b) Osteotomy at one site in the distal diaphysis of radius.

c) Osteotomy at two sites in the diaphysis of radius and ulna.

d) Osteotomy distal to the synostosis mass.

e) Derotation osteotomy with the Ilizarov method.

## EXCISION OF SYNOSTOSIS AND INTERPOSITION OF VASCULARISED FASCIO-FAT GRAFT

Early reports of operative treatment of this condition describe restoration of motion by resection of the synostosis and interposition of fat or muscle. However, this procedure led to uniformly poor results. Dal Monte et al. released the synostosis in twelve patients by removing part of the radius that was proximal to the synostosis and, in some patients by using aponeurotic or muscle flap to

preserve the separation of radius and ulna. He reported recurrence of synostotic bridge in every patient. Miura et al. operated on eight upper extremities in seven patients. They placed the anconeus between the radius and ulna, but the synostosis recurred in every patient.

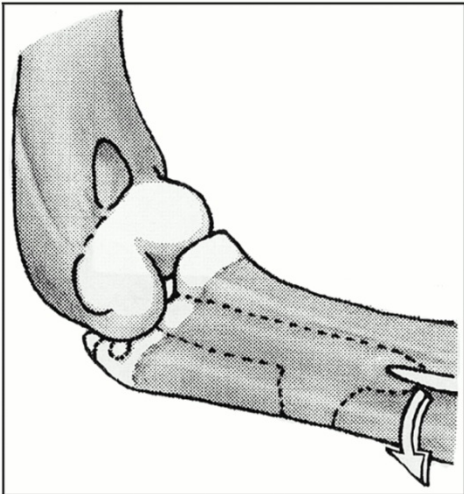
The excision of the synostosis can be done through a standard anterior Henry approach. Sometimes a posterior approach along the subcutaneous border of ulna is also necessary to adequately remove the synostosis and allow maximum passive range of motion.

The vascularized fascio-fat graft may be elevated from the mid-forearm posteriorly on the posterior interosseous vessels, with the pedicle graft being turned proximally and interposed into the synostosis site. However, the posterior interosseous artery may be anomalous for use in congenital synostosis. Kanya and Ibaraki (1) have used a free fascio-fat graft from the lateral arm with anastomosis of the posterior radial collateral vessels to available vessels in the proximal forearm. If the radial head is dislocated, a shortening angulation osteotomy is performed in the proximal radius. This osteotomy can be held with a four-hole dynamic compression plate.

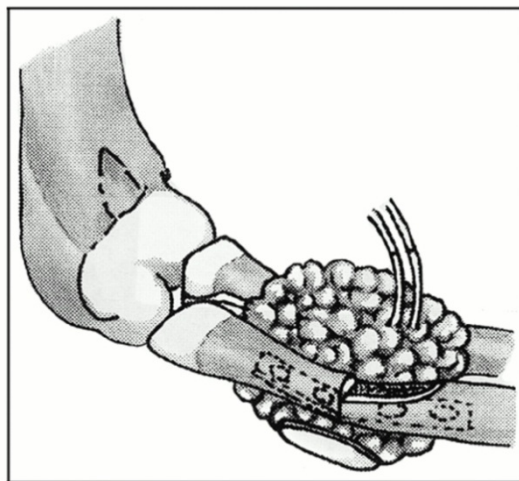
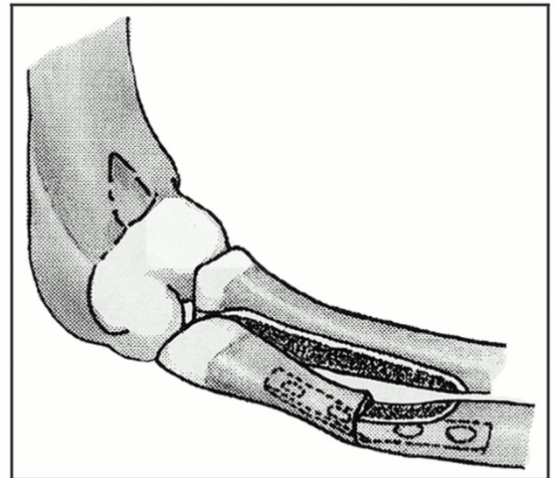


Free vascularised fascio-fat grafting described by Kanaya in 1998

Resection of synostosis



Radial shortening and plating



Interposition of vascularised fascio-fat graft

### Step 1: Separation and excision of synostosis

The synostosis is separated through an anterior and posterior approach. Posterior approach is first performed through an incision starting from the lateral epicondyle and curving dorsally to reach the posterior crest of ulna. The proximal and distal borders of the synostosis are identified with the use of injection needles. Synostosis is separated using a steel burr on a high-speed drill. Radial head is shaved to achieve forearm rotation. An anterior approach is performed and the biceps tendon is detached from the ulna to improve forearm rotation and to expose the space created by separation of synostosis.

### Step 2 : Radial shortening and plating

The radius is sectioned between the insertion of the pronator teres and the supinator, and the radial head is then reduced. The osteotomized ends of the radius is overlapped during reduction and the osteotomy is stabilized with a four-holed plate.

### Step 3: Interposition of vascularized fascio-fat graft

The vascularized fascio-fat graft, which included a small skin flap for use in monitoring the viability of the flap, is obtained from the ipsilateral arm. The graft is placed in the space created by separation of synostosis in a volar-to-dorsal

direction. The profunda humeri (donor) vessels are anastomosed with the recurrent radial (recipient) vessels. Wound is closed in layers after the sufficiency of the blood flow to the flap has been confirmed.

## DEROTATIONAL OSTEOTOMY OF THE PROXIMAL RADIUS AND DISTAL ULNA

Nguyen Ngoc Hung performed derotational osteotomy of the proximal radius and distal ulna in 39 patients (60 forearms). He recommended that this is a simple and safe technique to derotate the forearms of the patients with congenital radioulnar synostosis that are fixed in pronation.

In this method an osteotomy is done at two sites, one at the proximal radius shaft and the other at the distal ulna and a segment of bone is resected. K-wires, 1.5mm in diameter are passed into the radial and ulnar styloid, which are passed through the resecting bone sites. The forearm is then derotated manually into desired position.

Complications such as non-union, angulation of forearm, compartment syndrome, nerve palsy and vascular compromise were encountered in this study.

## DEROTATION OSTEOTOMY DISTAL TO SYNOSTOSIS MASS

M.Farzan, Kh.Daneshjou and S.M.J.Mortazavi performed a derotation osteotomy of the ulna distal to the synostosis mass in three patients. In this method, through a posterior approach the synostosis mass was reached and an osteotomy of ulna was done distal to the fusion mass. The limb was then derotated into desired after fixing the osteotomy site with a K-wire.

Vascular compromise, compartment syndrome, loss of derotation and loss of elbow range of motion were the complications.

## DEROTATION OSTEOTOMY THROUGH THE SYNOSTOSIS MASS

At present, the preferred surgical procedure is derotation osteotomy through the synostosis mass itself. Derotation osteotomy through the fusion mass appears to have many advantages as a method of placing the hand in a better functional position. The procedure is less difficult than attempting to change rotation by osteotomy at other levels and it preserves good coaptation of the divided ends. The osteotomy site, also heals rapidly.

William T. Green, MohinderA. Mittal performed transverse osteotomy at the site of the synostosis mass in thirteen patients. They obtained good results by this method and concluded that this is a safe, easy and effective method in treating

congenital radioulnar synostosis and also the complication rate associated with this procedure was much less when compared with other techniques.

## IDEAL POSITION OF THE FOREARM AFTER SURGICAL CORRECTION OF CONGENITAL RADIO-ULNAR SYNOSTOSIS

The ideal position to place the forearm after surgical correction remains controversial. The ideal position depends upon whether the deformity is unilateral or bilateral, dominant or non-dominant side, social and cultural environment of the patient and the projected future activities.

Green and Mittal suggested that in bilateral cases the best position was in 30° to 45° of pronation in the dominant forearm and in 20° to 35° of supination in the non-dominant. In unilateral cases, the ideal position was 10° to 20° of supination (4). Other authors have advocated 0° to 20° of supination in the non-dominant forearm and 0° to 20° of pronation in the dominant forearm.

### Recommended positions after osteotomy

Author	Unilateral	Bilateral	
		Dominant	Non dominant
Simmons	Neutral to 20 deg pronation	10-20 deg.pronation	Neutral
Green	10-20 deg. supination	30-45 deg.pronation	20-35deg.supination

### RECOMMENDED POSITION OF FOREARM IN INDIAN POPULATON

In bilateral cases

Right : 20-30 degrees of supination

Left : neutral to 20 degrees of pronation

In unilateral cases

Dominant hand in 20-30 degrees of supination

## **MATERIALS AND METHODS**

In our institution, 12 patients with congenital radioulnar synostosis were selected for this prospective study, after obtaining Ethical Committee approval.

All patients were treated with osteotomy through the synostosis mass and were followed up for a period of one year. The age group of the patients varied between 4-11 years, with the average being 6 years. The period of this prospective study was from July 2009-July 2011.

Of the 12 patients selected, 7 were male and 5 were female. 9 cases had bilateral radioulnar synostosis and 3 had unilateral radioulnar synostosis.

There was no family history of radioulnar synostosis in any of the patients and there was no syndromic association also in any of the cases.

### **INCLUSION CRITERIA**

- 1) Greater than 20 degrees of pronation on the right side with significant difficulties in daily activities, particularly eating.

### **PREOPERATIVE PLANNING**

X- ray of the involved upper extremity, both anteroposterior and lateral views were taken in all patients.

Preoperatively the position of ankylosis of the involved elbow joint and the range of supination and pronation of the involved upper extremity were measured. The mean preoperative degree of deformity was 45 degrees of pronation and the range being 20-60 degrees.

## ANAESTHESIA AND POSITION OF THE PATIENT

Surgery was done in a standard radiolucent table with the patient in supine position, with the involved upper extremity resting on a forearm table. All cases were done under general anaesthesia.

## SURGICAL TECHNIQUE

A longitudinal dorsal incision is made just to the radial side of the posterior aspect of the ulnar ridge, extending from a finger breadth distal to the olecranon tip distally for seven and one-half centimeters, depending on the size of the patient. Subperiosteal exposure of the fusion mass is easily accomplished. The site for the osteotomy is ordinarily at a comfortable level in the distal half of the fusion mass. The osteotomy line is marked out by fine drill holes made at intervals along the osteotomy site and an osteotome is used to complete the division. The bone is marked on either side of the osteotomy line so that the degree of rotation can be checked by the marks as well as by the position of the hand and forearm. After the



mass is divided, the distal mass is rotated into 20-30 degrees of supination. This is not difficult ordinarily, and it is comparatively easy to maintain good opposition of the fragments because they are cut transversely and are relatively broad. The diameter of the mass may be irregular enough so that in the corrected position removal of a little excess bone, if one protrudes over the other, may be desirable.

Two central pins are inserted from the olecranon distally across the osteotomy site, with the fragments in the desired position of rotation. The transverse pin is introduced with the forearm held in the desired position of rotation so as to minimize the stress on the soft tissue. The advantage of this method is that the longitudinal pin serves as a guide if the rotation needs to be changed and the transverse pin can be used to modify and hold the degree of rotation. However, in cases where the correction of deformity was easy the transverse was not introduced. A long arm cast is applied with the elbow in 90 degrees of flexion and the hand supported just proximal to the metacarpal heads.

Radial pulse and the circulation of the thumb are assessed as the deformity is corrected, before final closure and regularly in the immediate post-operative period for evidence of any possible impairment. If any difficulty arises in the circulation, it is most likely to be associated with the degree of rotation. It is desirable,

therefore, to be able to derotate the correction even temporarily should there be any evidence of circulatory or neural difficulty.

## POST-OPERATIVE PROTOCOL

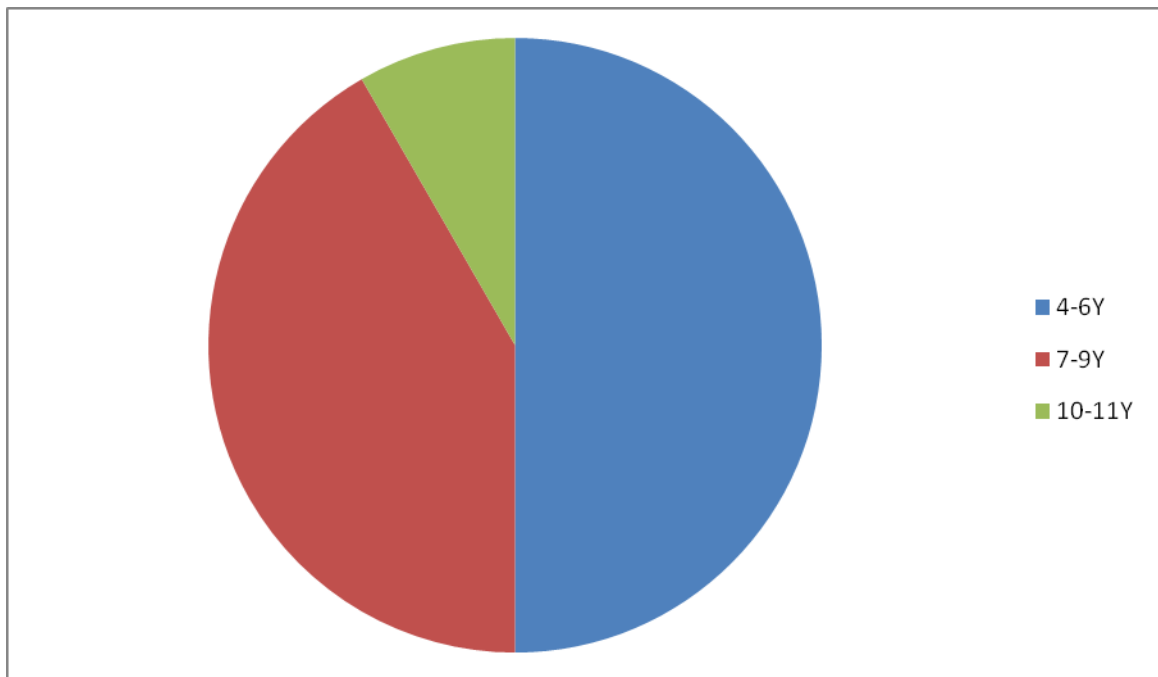
The upper limb must be immobilized with a long arm cast with the elbow in 90 degrees of flexion in the immediate post-operative period. The limb is immobilized for a period of 8 weeks during which serial radiographs are taken to assess bony union. Once bony union is achieved radiologically, active mobilization of the elbow joint can be started.

## OBSERVATIONS AND RESULTS

### AGE GROUP

AGE IN YEARS	NO OF PATIENTS
4-6	6
7-9	5
10-11	1

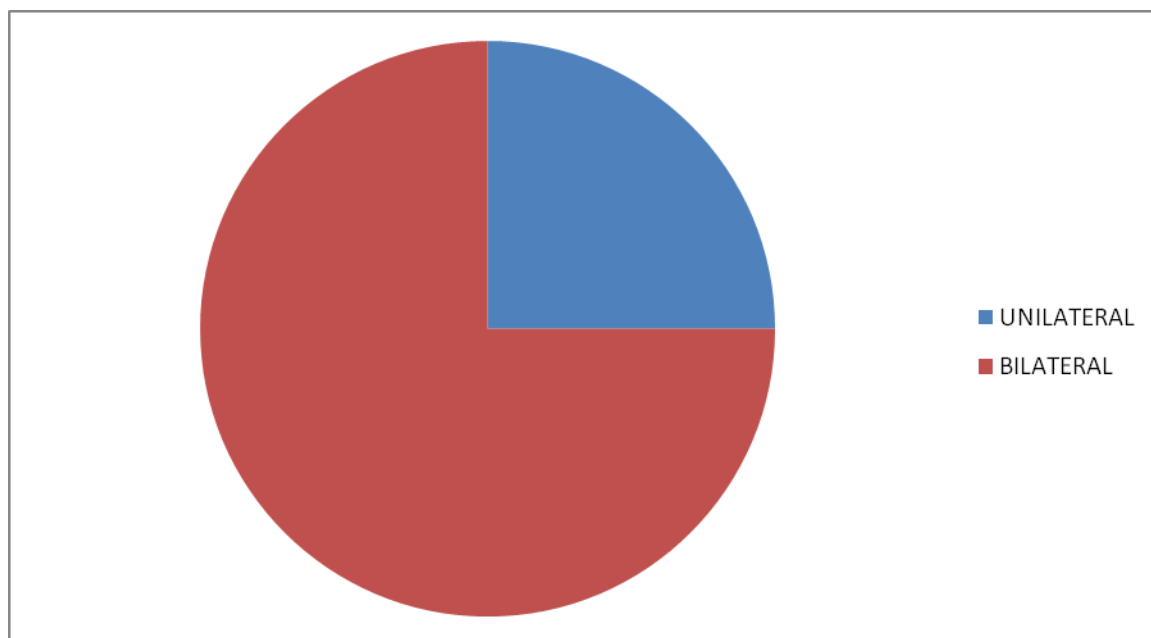
Total of 12 cases were selected for our study, out of which 6 patients were in the age group of 4 to 6 years, 5 were in the age group of 7 to 9 years and 1 was in the age group of 10 to 11 years.



## LATERALITY

LATERALITY	NO OF PATIENTS
UNILATERAL	3
BILATERAL	9

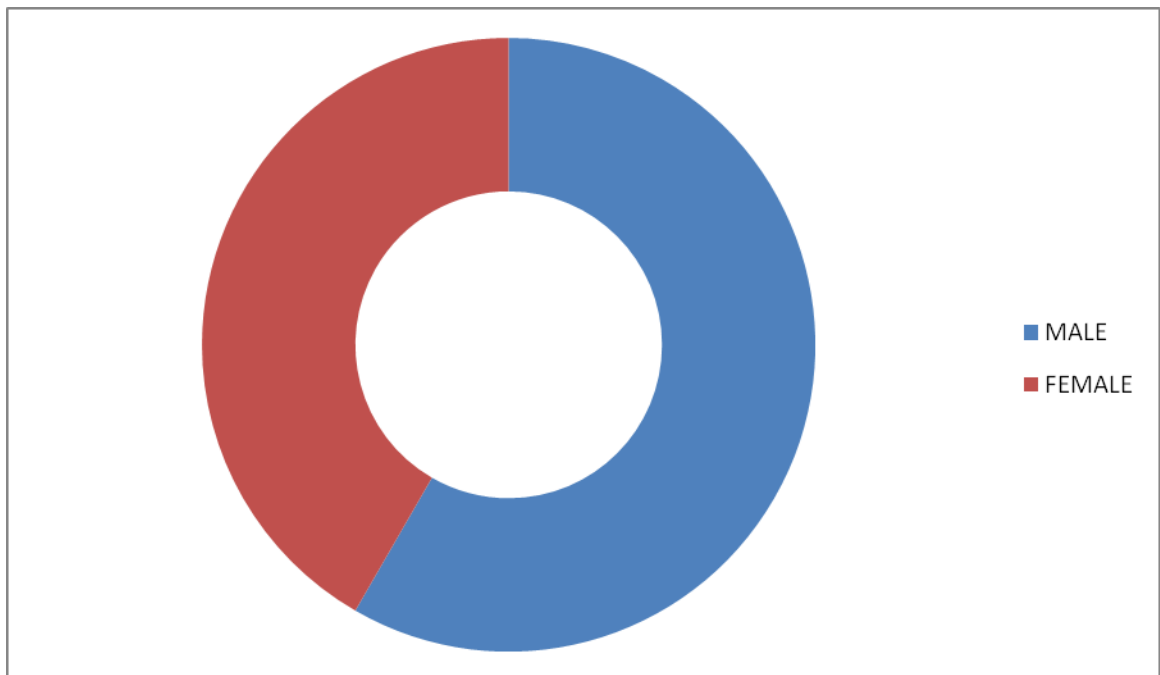
9 cases had bilateral radioulnar synostosis and 3 patients had unilateral radioulnar synostosis.



## SEX RATIO

SEX	NO OF PATIENTS
MALE	7
FEMALE	5

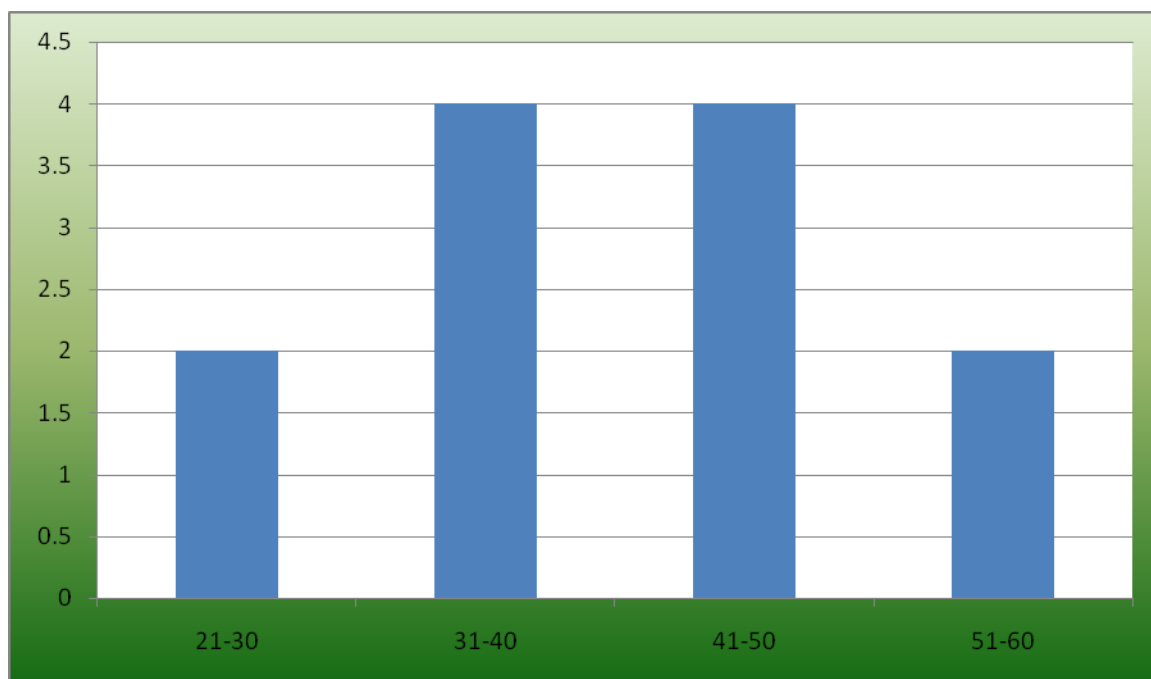
Out of the 12 patients, 7 were male and 5 were female.



## PRE-OPERATIVE DEFORMITY

DEFORMITY(IN DEGREES OF PRONATION)	NO OF PATIENTS
21-30	2
31-40	4
41-50	4
51-60	2

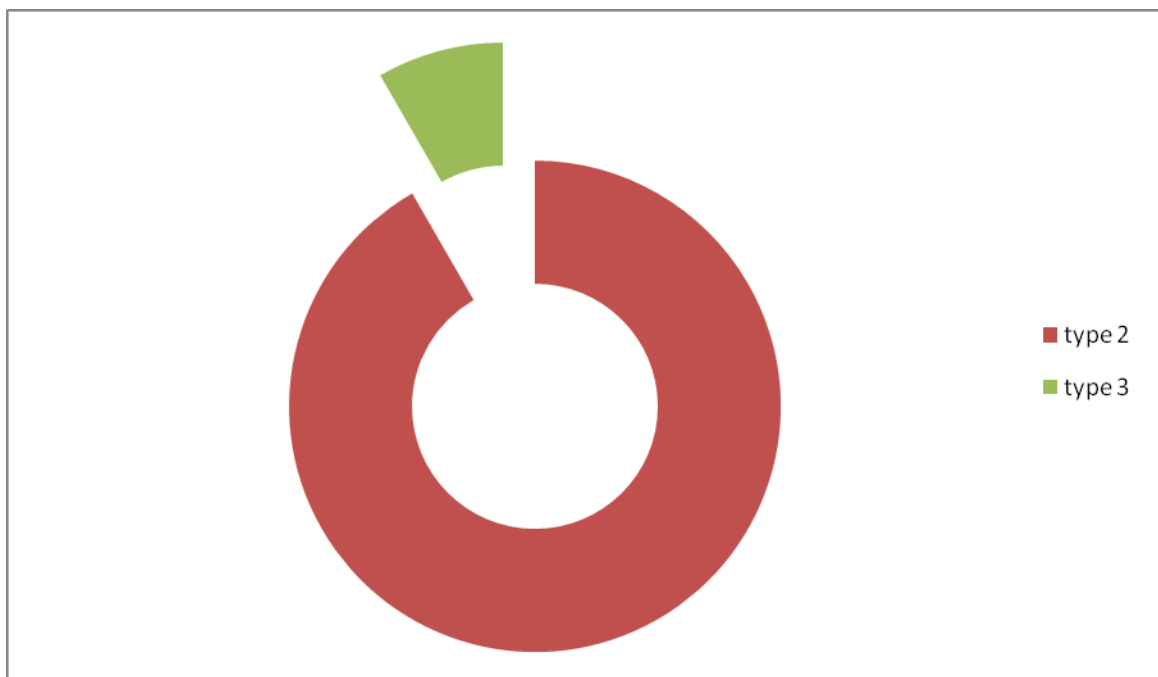
2 patients had severe deformity with the forearm being in 50 to 60 degrees of pronation.



## TYPE OF SYNOSTOSIS

Cleary and Omer type	No of patients
Type 1	-
Type 2	11
Type 3	1
Type 4	-

The most common type of synostosis encountered in our study was Cleary and Omer type 2 synostosis.



## CASE ILLUSTRATION

### CASE 1

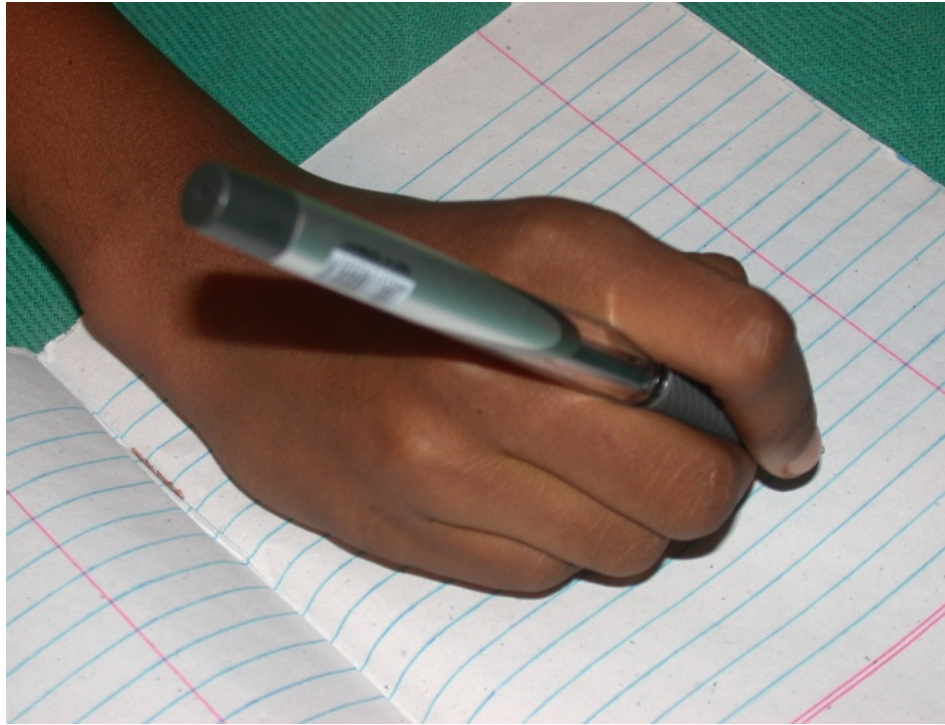
6 years old female patient of type II Radioulnar synostosis on the right side with complaints of difficulty in eating and writing. Preoperative deformity was found to be 50 degrees of pronation on the right side.

Picture showing difficulty in eating with preoperative deformity of 50 degrees of pronation





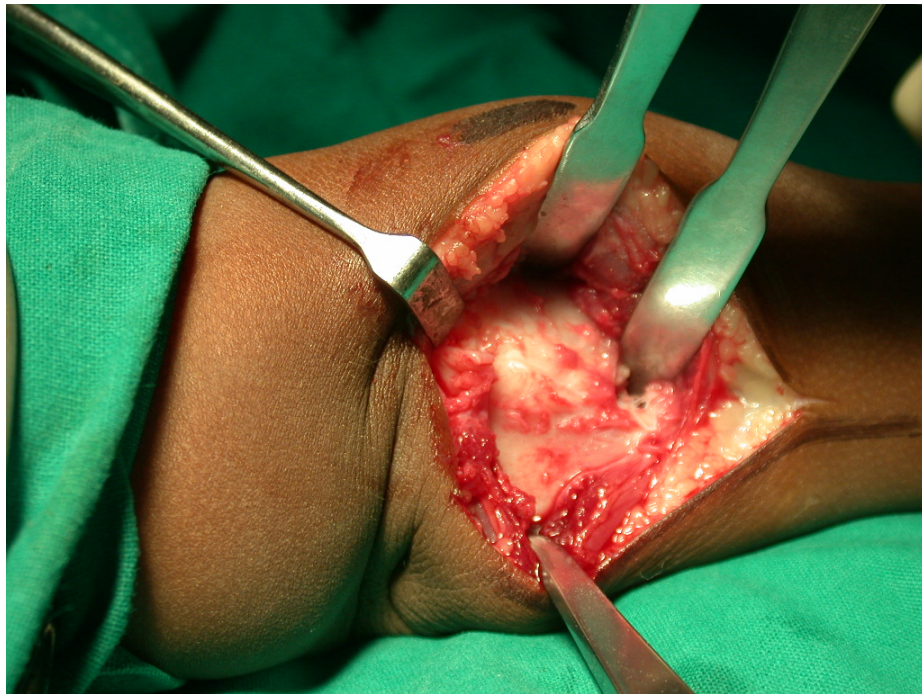
Picture showing difficulty in writing



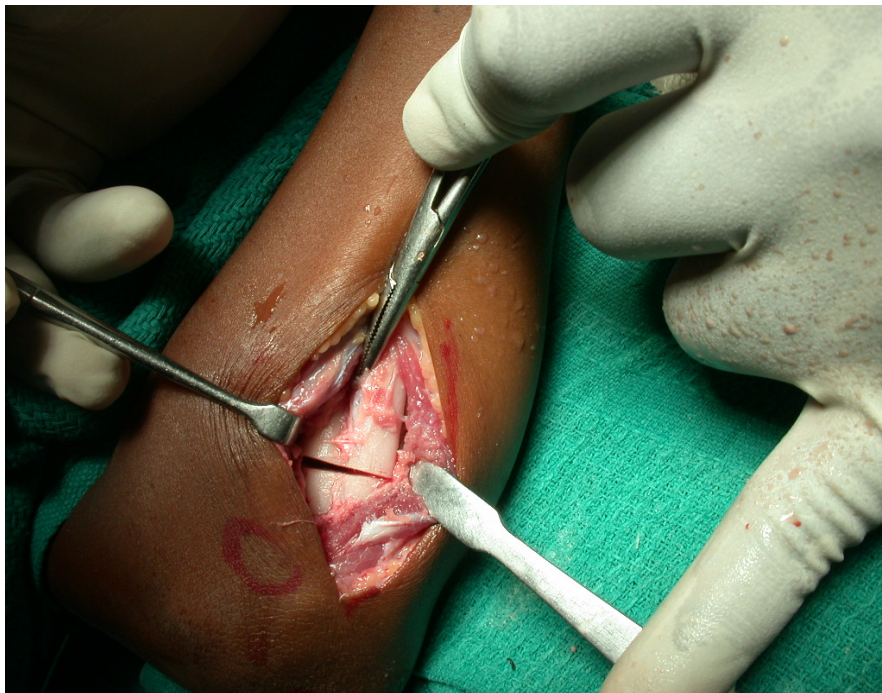
Type 2 radioulnar synostosis



Exposure of the synostosis mass



Transverse osteotomy through the synostosis mass



Post operative x-ray showing k-wire fixation



8 weeks post operative x-ray showing signs of union

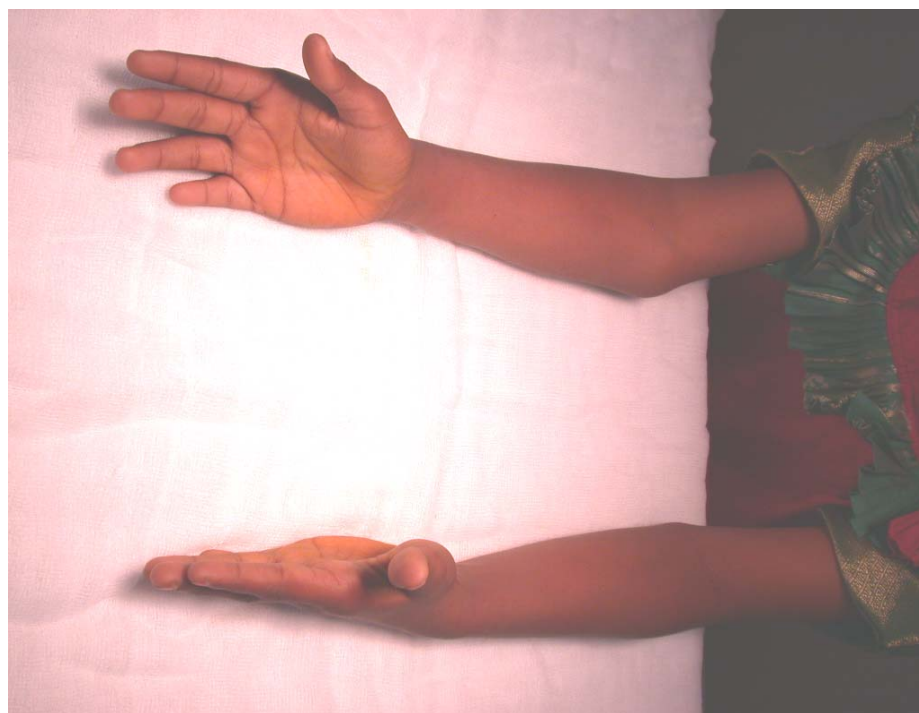




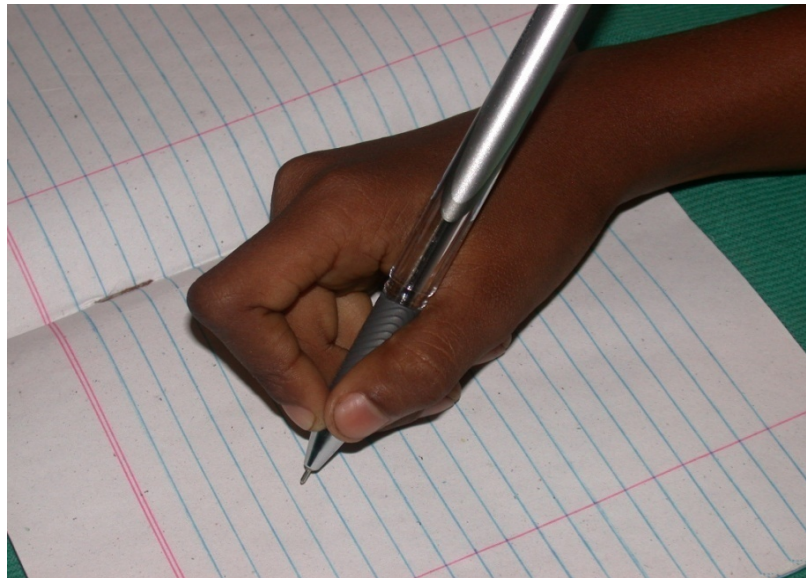
Preoperative deformity in 50 degrees of pronation on the right side



Post operative position of 30 degrees of supination on the right side



Improvement in ability to write



Improvement in ability to eat



## CASE 2

4 years old male patient of type II Radioulnar synostosis on the right side with complaints of difficulty in eating and writing with preoperative deformity of 40 degrees of pronation.

Difficulty in eating

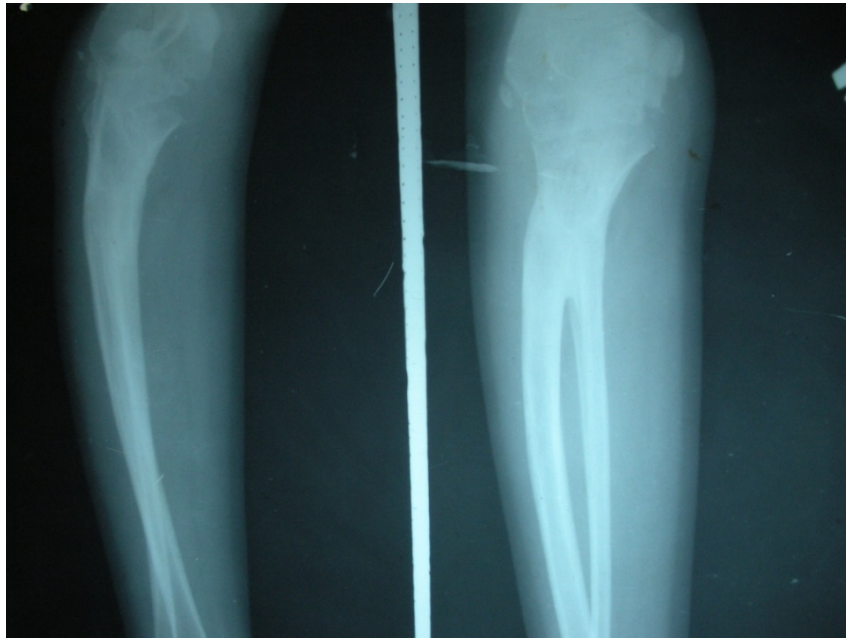


Inability to supinate

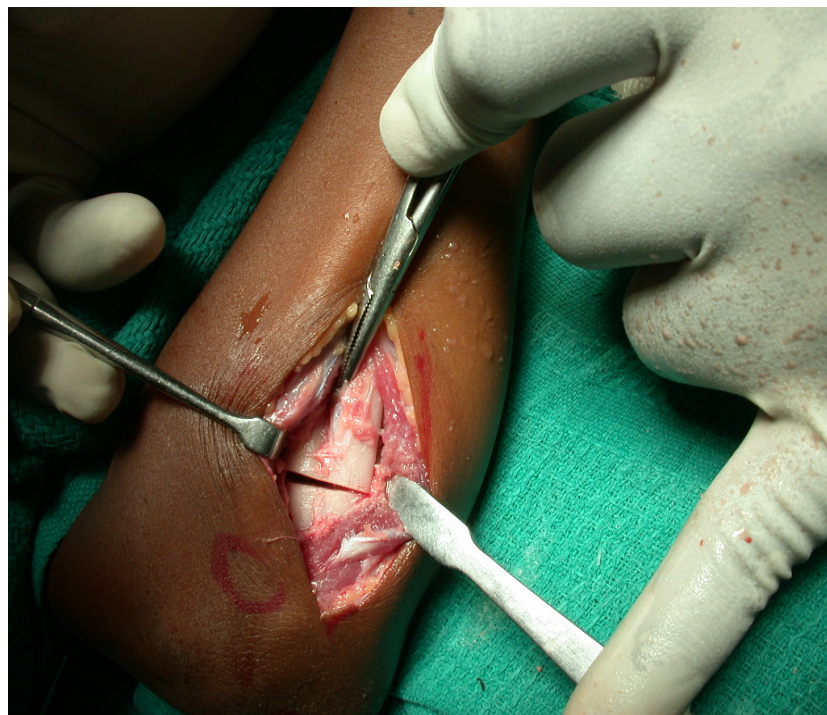




## Type 2 radioulnar synostosis



## Osteotomy through the synostosis mass



## Osteotomy and k-wire fixation



8 weeks postoperative picture showing signs of union





Postoperative position of 25 degrees of supination



Improvement in ability to eat



CASE 3

POSTOPERATIVE PICTURES



Position of right forearm-30 degrees of  
supination



Normal eating posture



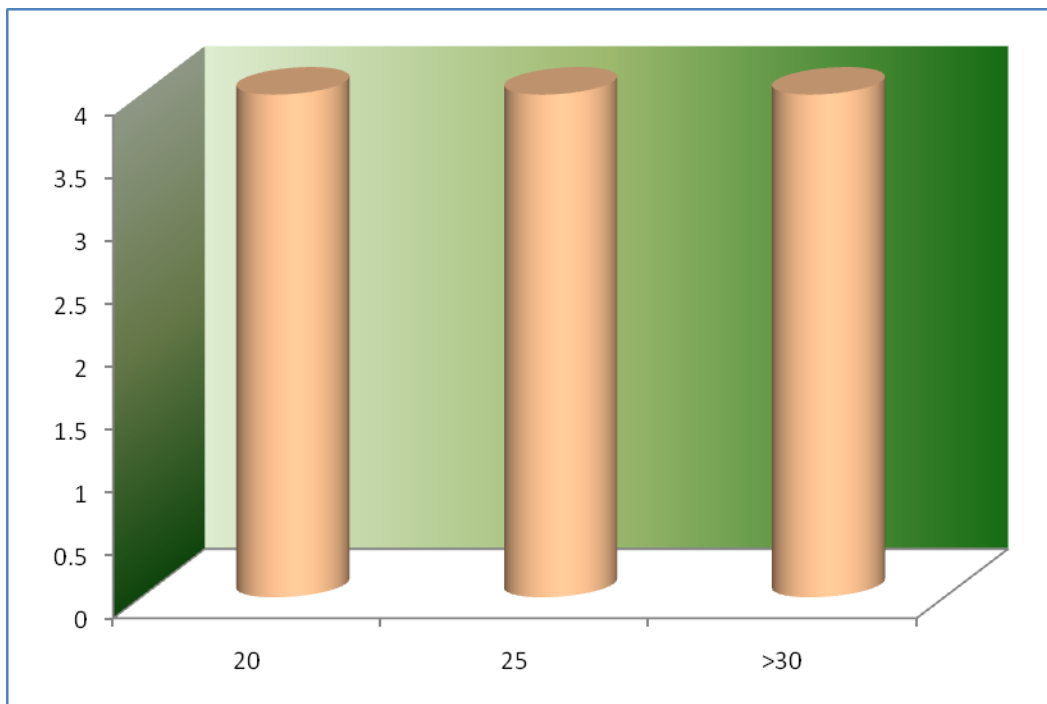
Ability to receive objects in open palm



Improvement in ability to write

## POST-OPERATIVE POSITION OF FOREARM

FOREARM POSITION IN DEGREES OF SUPINATION	NO OF PATIENTS
20	4
25	4
>30	4



## RESULTS

The length of the follow-up for these patients was one year. Evaluation of the results was based on the following parameters: the position of the forearm, appearance, function, and the patient's and family's opinion of the patient's status in comparison with the previous preoperative condition. At the end result examination, four forearms were in 20-25 degrees of supination, four were in 26-30 degrees of supination and four in supination greater than 30 degrees. There was no loss of correction in any of the cases.

All of the cases were rated as improved and all but one had a smooth post-operative course. One patient had transient posterior interosseous nerve palsy, which recovered in three weeks time with conservative line of management.

The results were graded as excellent in four patients (forearms in supination greater than 30 degrees) in whom anatomical and functional results were such that they had no handicap in normal activities and the position of the forearm was considered ideal. The results in eight patients were graded as good, functionally, as the patients would have preferred 10-20 degrees more supination on the forearm operated on. Bony union was achieved at the end of 8 weeks in all the 12 cases.

## COMPLICATIONS

Other than the one complication of transient posterior interosseous nerve palsy which recovered in three weeks, no other vascular or neural complication was encountered. There were no cases of infection or wound dehiscence.

## **DISCUSSION**

Rotational osteotomy through the fusion mass appears to have many advantages as a method of placing the hand in a better functional position. The procedure is less difficult than attempting to change rotation by osteotomy at other levels and it preserves good coaptation of the divided ends. The osteotomy heals rapidly and only one complication occurred in our patients. As a great deal of rotatory correction is involved in the operation, the circulation must be carefully observed, and undue stress during the correction should be avoided. No untoward neural signs or symptoms occurred in our patients as a result of the change in position, but care must be taken that they do not arise.

Osteotomy through the forearm bone has many disadvantages such as greater soft tissue restriction, loss of correction during cast immobilization, risk of vascular compromise and in late childhood cases bony union may be delayed. The main advantage of our technique being its simplicity and safety, use of single incision, less soft tissue restriction during correction, comparatively easy k-wire fixation, achievement of union in all cases and absence of severe postoperative complications.

Several authors have reported separation of the synostosis and the interposition of fat or muscle (or some other material), but recurrence of the ankylosis has still been noted. Kelikian and Doumanian reported good results with the use of swivel prosthesis in patients who had post-traumatic proximal radioulnar synostosis; however, Tachdjian noted disappointing results with the use of swivel prosthesis in patients who had congenital synostosis, with recurrence of the ankylosis in eighteen months follow-up (37).

Simmons et al. (6) reported eight complications in association with twenty two osteotomies; the complications included a wound infection (one), loss of correction (three), and circulatory problems (four). Green and Mital (4) reported one ischaemic contracture in thirteen patients. Wide exposure and careful protection of the neurovascular structures may be the reason why none of our patients had neurovascular complications.

The ideal position to place the forearm after surgical correction still remains controversial. An important factor entering into this choice of the position of the forearm and hand is the relation of the motion of the shoulder to a hand that does not have the benefit of motion between the radius and the ulna. A hand in a position of supination can do most things that require supination and yet, when in a forward working position with a flexed elbow, it can attain a degree of functional

pronation by controlled amounts of internal rotation, flexion, and abduction at the shoulder. Conversely, if the hand is in pronation in the forward working position, substitute motions of the shoulder are of little help in placing the hand in a position of functional supination. Unless there is some degree of supination in one hand, there is great difficulty in doing gracefully many of the things that are a part of daily activity, such as receiving change in coin. If one hand is in a supinated position it complements the other, which is in pronation, in every way including palmar apposition, so that even if the other hand is not corrected the patient gets along very well with added mobility developed in the wrist. In patients with unilateral synostosis a corrective surgical procedure is less imperative unless the position of pronation is extreme. Function can be greatly improved in many arms, however, if the pronation is decreased to the desired position for the forearm, 10 to 20 degrees of supination. Similarly, it may be desirable to rotate both forearms in patients with bilateral synostosis, assuming that if one is placed in the desired position of supination, the other may need to be placed in 10 to 20 degrees of pronation to assure that the patient is able to perform tasks requiring supination and pronation.



## CONCLUSIONS

1. Congenital radioulnar synostosis is a rare deformity, frequently bilateral, and more commonly seen in male patients.
2. The etiology is multifactorial , with both sporadic mutations and undefined genetic patterns.
3. Surgical indications are based on the degree of deformity and bilaterality.
4. If bones of the forearm are in ideal position of rotation, the use of both hands can be amazingly efficient without the motions of supination and pronation.
5. From our study, we conclude that if the position of the forearm needs to be changed, a well controlled rotational osteotomy through the conjoined radio-ulnar mass is the best way of accomplishing this.

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# **PROFORMA**

**Name:**

**Age:**

**Sex:**

**Address:**

**IP No:**

**Unit:**

**DOA:**

**DOS:**

**Ward:**

**Unilateral/bilateral :**

**Side of synostosis :**

**Cleary and Omer Classification:**

**Any associated anomaly :**

**Investigations:**

- Plain X-ray elbow joint with forearm- AP and lateral view
- Blood Hb/Sugar/ Urea/ Creatinine/ Grouping and Typing



- Chest X-ray

### **Surgery:**

- Type of anaesthesia
- Patient positioning
- Approach
- Operative time

### **Post Operative:**

- Mobilisation
- Type of physiotherapy

### **Complications:**

- Infection- Early/Late
- Vascular compromise
- Nerve injury
- Loss of correction
- Non union

### **Follow Up:**

- Radiological Assessment: X-ray elbow joint with forearm , AP and lateral view at 3 wks, 6wks and 8 wks.

# Master Chart

Case	Age at Surgery / Sex	Involved side	Position of ankylosis		Preop elbow ROM	Shortening of I/ L Extremity	Wrist hypermobility	Cleary and Omer type	Post- op results	
			L	R					Position of Forearm®	Complications
1	5y/f	Bilateral	20 pro	50 pro	full	No	No	ii	20sup	None
2	6y/f	bilateral	10 pro	50 pro	Full	No	Yes	li	20Sup	None
3	8y/f	Right	-	50 Pro	Full	No	No	li	25Sup	None
4	5y/m	Bilateral	25 pro	35 pro	Full	No	No	li	20Sup	None
5	5y/f	Bilateral	20 Pro	55 Pro	Full	No	No	li	25Sup	Transient pin palsy
6	6y/m	Bilateral	30 pro	60 Pro	Full	No	No	li	>30Sup	None
7	6y/f	Bilateral	35 pro	40 Pro	Full	No	No	li	>30 Sup	None
8	7y/m	Right	-	50 Pro	Full	No	Yes	li	20Sup	None
9	9y/m	Right	-	35 Pro	full	No	No	li	25Sup	None
10	11y/m	Bilateral	10 pro	30 pro	Full	no	no	ii	25sup	None
11	7y/m	Bilateral	20 pro	40 pro	Full	no	no	ii	>30 sup	None
12	8y/m	Bilateral	20 pro	30 pro	Full	no	no	ii	>30 sup	None

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